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Syringomyelia with type I Arnold-Chiari malformation

Syringomyelia is a heterogenous group of conditions characterized by the presence of abnormal neuroglial cavities (syrinx) filled with cerebrospinal fluid, within the spinal cord or truncus cerebri. The usually irregular and longitudinal spinal cavity is located in the midline of the cord, beginning in the cervical part and can extend as far as the caudal part (even all the way along the spinal cord) (6). Syringomyelia is most commonly associated with Arnold–Chiari I malformations (this is a congenital anomaly in which the cerebellar tonsils are displaced downward through the foramen magnum into the upper cervical spinal canal) (3, 7). Sometimes it can also be seen with arachnoiditis , intraspinal tumors or intraspinal infection (2).

The etiology of syringomyelia is not clear. The Hydrodynamic Theory of Gardner (1965) is based on the "water-hammer" pulse effect of the cerebrospinal fluid, which forced into the central canal of the spinal cord, pushes it outward, causing syrinx formation. Williams (1981) attributes the major role to increased intracranial pressure caused by flow disturbances within foramen magnum region. Oldfield (1994) championed the "piston effect" of the herniated cerebellar tonsils. The latest theory of Levin integrates both William's and Oldfield's theories and emphasizes "mechanical stress" as the main factor causing changes in cerebrospinal fluid pressure leading to spinal tissue damage (4, 8).

Syringomyelia may be asymptomatic or symptomatic. The main symptom is dissociated sensory loss (pain and temperature loss with preservation of light and touch perception and proprioceptions). Weakness of upper extremities with hand muscle atrophy may occur (5, 6). Sometimes syringomyelia is a cause of body hypertrophy (9).

The most common symptoms of type I Arnold-Chiari malformation are suboccipital headache that is aggravated by coughing, straining, physical exertion, Valsalva maneuvers, and weakness – especially in the hands. Other signs include neck pain, arm pain, numbness, loss of temperature perception, ataxia, diplopia, dysarthria, vomiting, vertigo, nystagmus, tinnitus (3, 7). Magnetic resonance image of the spinal cord and foramen magnum region makes it possible to detect syringomyelia and Arnold-Chiari malformation. The presence of type I Arnold-Chiari type I malformation is an indication for the need of computer tomography examination to estimate ventricle system (1).

This work describes a clinical case of a patient hospitalized due to syringomyelia associated with asymptomatic type I Arnold-Chiari syndrome.

CASE DESCRIPTION

A 41-year-old female patient was admitted to the Neurology Ward in a hospital in Lublin due to pain in the left shoulder which lasted for 6 months, and parestesia in the left upper limb lasting for 4 months which was associated recently with a burning pain sensation. Two weeks before hospital admission, numbness of left leg occurred. There were no other complaints registered. Her medical history records 2 cesarean operations, and one child born with upper lip cleavage defect.

The physical investigation showed two scars of diameter about 14 mm, which occurred after recent burns and a decreased temperature in both left limbs. The neurological investigation showed

partial dysfunction of trigeminal nerve within all three branches. Movements of all limbs were not restricted. The perception of pain, temperature and superficial sensibility was considerably decreased within left limbs. The muscle strength or profundi reflexes do not show differences between both sides.

MRI scans of cervical and thoracic regions of the spine revealed large cavity filled with cerebrospinal fluid which extends from C3 to Th7 with signs of tonsillar herniation into the foramen magnum (Fig. 1). The X-ray picture of the occipito-altoid junction did not show any changes in the bone structure in this region. To estimate the brain chamber system, a CT head examination was ordered, which did not reveal hydrocephalus, oedema nor any other significant changes of the brain structures. Based on the neurological examination as well as on MRI pictures of the spinal cord, a diagnosis was made: syringomyelia with Arnold-Chiari type I syndrome.

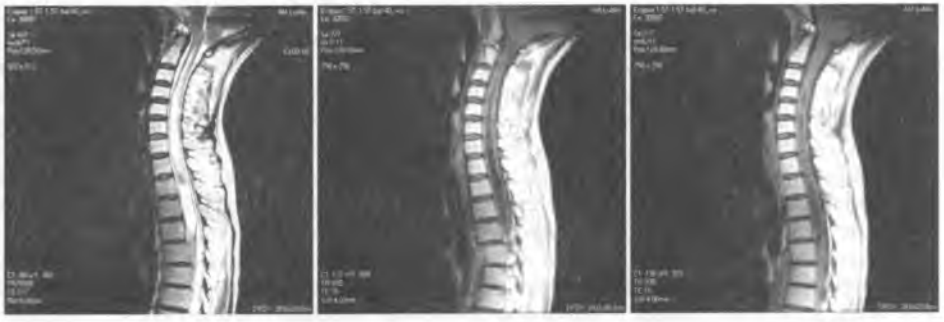

Fig. 1. The described case in T2 and T1 MRI scans

DISCUSSION

Syringomyelia appears usually in young adulthood and progresses very slowly. The symptoms mostly depend on syrinx localization (6). The dissociated sensory loss is often a cause of burns and associated scars. In this particular case they were the result of the thalamo–spinally path damage. Other common symptoms such as hand-muscle atrophy (damage of spinal corni anterior), Horner's symptom (damage of ciliospinal center), and spastic paresis of lower extremities (damage of pyramidal tract), were not observed.

Arnold-Chiari syndrome belongs to the family of dysraphic anomalies connected with central tube closing disorders. Chiari malformation type I is caused by the displacement of cerebellar tonsils down below the occipital opening. The clinical symptoms result from the pressure on medulla oblongata or from the hydrocephalus presence (3, 7). In this particular case, typical Arnold-Chiari symptoms were not observed, except for a burning pain in the left arm. The presence of Arnold-Chiari symptoms is an indication for the need of MRI or CT examination to assess the cerebrospinal fluid circulation and ventricular system condition (1), yet a CT scan did not show any abnormalities within this system, or any features of hydrocephalus.

CONCLUSION

The presented case of a patient with diagnosed syringomyelia along with Arnold-Chiari type I syndrome leads to the conclusion that these disease entities may be revealed late in life or can exist with very mild symptoms.

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SUMMARY

We present the case of syringomyelia with Arnold-Chiari malformation in a female patient who was having a pain and parestesia in the left upper limb (the perception of pain, temperature and superficial sensibility was considerably decreased within left limbs). MRI scans of cervical and thoracic regions of the spine revealed large cavity extends from C3 to Th7 with signs of tonsillar herniation into the foramen magnum.

Jamistość rdzenia z zespołem Arnolda-Chiariego typu I

Przedstawiono przypadek jamistości rdzenia z zespołem Arnolda-Chiariego u pacjentki z bólem i parestezjami w lewej kończynie górnej, z osłabieniem czucia bólu, temperatury, czucia powierzchownego w lewych kończynach. MRI odcinka szyjnego i piersiowego kręgosłupa wykazało dużą jamę rozciągającą się od C3 do Th7 oraz wklinowanie migdałków móżdżku do otworu potylicznego wielkiego.